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BRAIN PATHOLOGY CASE OF THE MONTH MAY 2019 - CASE 1

69-YEAR-OLD MALE WITH AN INTRADURAL, EXTRAMEDULLARY MASS AT T12-L1

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CLINICAL HISTORY

In 2002, a 55-year-old male patient was referred for neurosurgical consultation after two episodes of speech arrest and a newly diagnosed mass lesion in the superior left temporal gyrus extending to the insula. The patient underwent a pterional craniotomy with partial resection of the tumor. The histopathological examination revealed an oligodendroglioma (WHO II). A follow-up MRI in 2007 demonstrated significant growth of the residual tumor mass (Figure 1A–D). Accordingly, the patient was started on standard therapy with temozolomide. In 2011, MRI showed tumor progression, which prompted additional tumor resection. The histological examination revealed features of anaplastic oligodendroglioma with codeletion of 1p/19q. Postoperative treatment included adjuvant radiotherapy (32 × 1.8 Gy = 57.6 Gy). In June 2015, follow-up MRI revealed a local recurrence consistent with tumor progression. Meanwhile, the patient had undergone surgical resection of adenocarcinoma of the colon, diagnosed in 2015, and treated with adjuvant chemotherapy (FOLFOX4). The decision was made to suspend further treatment and to follow the patient with MRI. Of note, the regression of the tumor mass was observed in the parahippocampal gyrus.

In September 2016, 14 years after the initial diagnosis of oligodendroglioma, follow-up MRI of the liver was performed according to the staging protocol for metastatic adenocarcinoma. MRI of the abdomen revealed the incidental finding of an intradural tumor attached to the conus medullaris. MRI of the lumbar spine showed a circumscribed, intradural and extramedullary tumor (11 × 13 × 24 mm). The tumor occupied the spinal canal between Th12-L1, shifting the conus to the left, as well as infiltrating the fibers of the cauda equina (Figure 1E–H). The clinical examination revealed mild paresis of the right leg, in particular, the right hip and foot flexors (M4/5).

Laminoplasty was performed at Th12-L1 and the intradural and intramedullary lesion were resected. The postoperative course was uneventful and the patient did not show any new deficits after surgery (Figure 1I–L, postoperative MRI). The histological diagnosis is discussed below in detail.

MICROSCOPIC PATHOLOGY

Multiple tissue fragments measuring 8 × 8 × 1 mm in total were submitted for the histological examination. The microscopic examination revealed a neoplasm composed of closely packed and relatively uniform round cells with perinuclear halos. Frequent mitoses were observed, which were accompanied by microvascular proliferation and pseudopalisading necrosis (Figure 1M–O). Scattered calcospherites were present. The nuclei and cytoplasm of the tumor cells were immunoreactive for the IDH1^{R132H}-mutation. ATRX immunohistochemistry showed retained nuclear expression. The tumor cells were epithelial membrane antigen (EMA) negative and glial fibrillary acidic protein (GFAP) positive (Figure 1P–Q). The MIB1-proliferation index was high, reaching 50% (Figure 1R). **What is your diagnosis?**

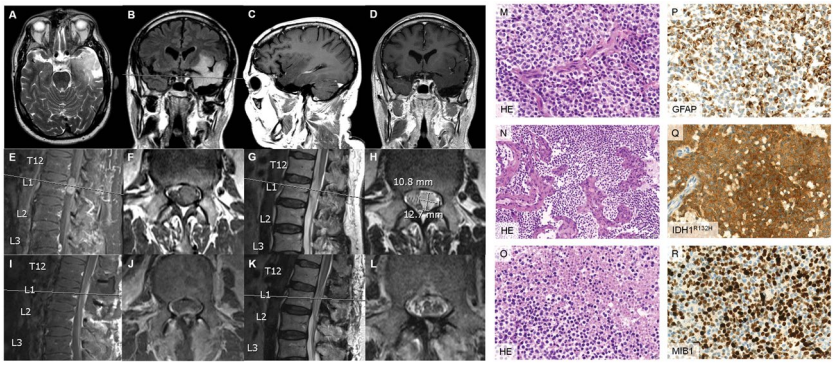

Figure 1.

DIAGNOSIS

Drop metastasis of an anaplastic oligodendroglioma.

DISCUSSION

Malignant progression in patients with oligodendroglioma is inevitable (3). However, the prognosis is generally more favorable than astrocytomas of the corresponding grade. Oligodendrogliomas, which usually arise in the cerebral hemispheres, primarily recur locally, typically within the resection margins or treatment fields (2). Nevertheless, spinal drop metastasis from oligodendroglioma is a rare finding with fewer than dozen cases since first reported in 1971 (1) In the present case, 14 years elapsed after the initial diagnosis of oligodendroglioma. During a staging MRI for metastatic adenocarcinoma, an intradural tumor attached to the conus medullaris was detected, which was considered compatible with schwannoma or ependymoma. Surprisingly, the histopathological examination revealed an anaplastic oligodendroglioma. Of note, additional tumor resection of the intracranial oligodendroglioma was performed four months after the resection of the spinal drop metastasis. The histological examination revealed features of anaplastic oligodendroglioma with codeletion of 1p/19q.

While primary brain tumors such as glioblastoma, followed by primitive neuroectodermal tumors (PNET) and ependymomas are more likely to metastasize to the spinal canal, metastatic spread of oligodendroglioma is exceedingly rare (1). The mechanism underlying the metastatic spread of oligodendroglioma is controversial, with possible pathways including tumor dissemination via the cerebrospinal fluid pathways or even hematogenous spread. In cases of leptomeningeal disease or scalp involvement associated with multiple craniotomies, spread along lymphatic channels to regional lymph nodes followed by distal metastases is discussed (2). Tumor dissemination via the cerebrospinal fluid, reported in up to 14% of patients with oligodendroglioma, is thought to arise after surgical intervention with breaching the pia mater and ependymal lining (4). Isolated intramedullary metastases are suggested to occur from hematogenous dissemination.

The short survival in patients with anaplastic oligodendroglioma combined with the relatively slow growth rate has been considered as an explanation for the rare occurrence of metastasis (1). Thus, in the future, the incidence of patients with spinal drop metastasis of oligodendroglioma is likely to increase due to improved therapies and longer survival of patients.

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